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Background of extreme weight loss and weakness – right atrial myxoma

Pozadina izrazitog gubitka težine i slabosti – desni atrijski miksom

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Summary

A 63-year-old man was admitted in hospital because of extreme weight loss (about 20 kg within 6 months), weakness, fatigue, exertional dyspnea, periods of hacking cough, and laboratory disturbances (elevated erythrocyte sedimentation rate, increased C-reactive protein values, disturbances in the protein electrophoresis), with no proper etiology. Before admission he was extensively examined due to the commonest reasons for the mentioned symptoms – occult malignancy, hematological, autoimmune, and infectious diseases. Since extensive diagnostic evaluation performed before hospitalization showed no discrepancies, we suspected a cardiac myxoma. Echocardiography revealed a large tumor in the right atrium. The histology of surgically removed tumor revealed a cardiac myxoma. Five months later, his body mass increased by 15 kg. The patient remains well, with normal echocardiographic and laboratory findings. Our case suggests that echocardiography should be considered early in the examination of patients with markedly constitutional symptoms and laboratory disturbances in order to detect rare but treatable heart tumors.

Ključne riječi: constitutional symptoms, echocardiography, myxoma, weight loss

Sažetak

63-godišnji muškarac hospitaliziran je zbog izrazitog gubitka tjelesne mase (oko 20 kg tijekom 6 mjeseci), slabosti, umora, nedostatka zraka tijekom tjelesnih aktivnosti, povremenog kašljucanja i laboratorijskih poremećaja (ubrzana sedimentacija eritrocita, povećane vrijednosti C-reaktivnog proteina, otkloni u vrijednostima elektrofereze proteina) nepoznatoga uzroka. Pred prijam opsežno je obrađen zbog najčešćih uzroka navedenih simptoma – malignih, hematoloških, autoimmunih i zaraznih bolesti. Budući da dijagnostička obrada provedena prije hospitalizacije nije pokazala odstupanja, posumnjali smo na miksom srca. Ehokardiografskim pregledom zabilježen je veliki tumor u desnom atriju. Histološkom analizom kirurški odstranjenog tumora potvrđen je miksom. Pet mjeseci nakon operacije bolesnik se osjećao dobro, tjelesna masa mu se povećala za 15 kg, a ehokardiografski i laboratorijski parametri bili su u normalnim rasponima. Naš slučaj pokazuje da ehokardiografiju treba primijeniti u ranoj fazi obrade bolesnika s izraženim konstitucionalnim simptomima i laboratorijskim poremećajima, u cilju otkrivanja rijetkih, ali izlječivih tumora srca.

Key words: konstitucionalni simptomi, ehokardiografija, miksom, gubitak težine

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Introduction

Primary cardiac neoplasms are rare entities among heart diseases with an estimated incidence ranging 0.0017-0.33%.[1,2] About three quarters of these tumors are benign; myxomas being the most prevalent.[1,2] Clinical suspicion and prudent interventions are required for early diagnosis, which is the key to reducing morbidity and mortality. However, nonspecific systemic (constitutional) symptoms, occasional cardiac and/or embolic phenomena may be overlooked in the absence of any history of cardiac diseases. Therefore, cardiac evaluation may not be performed or ignored, and the diagnosis of this uncommon condition may be

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delayed until the onset of a more significant disease, or even sudden death.[1,2]

We present a patient with extreme weight loss, weakness, fatigue, exertional dyspnea, hacking cough and laboratory abnormalities due to the massive right atrial myxoma who underwent surgical intervention with significant improvement.

Case presentation

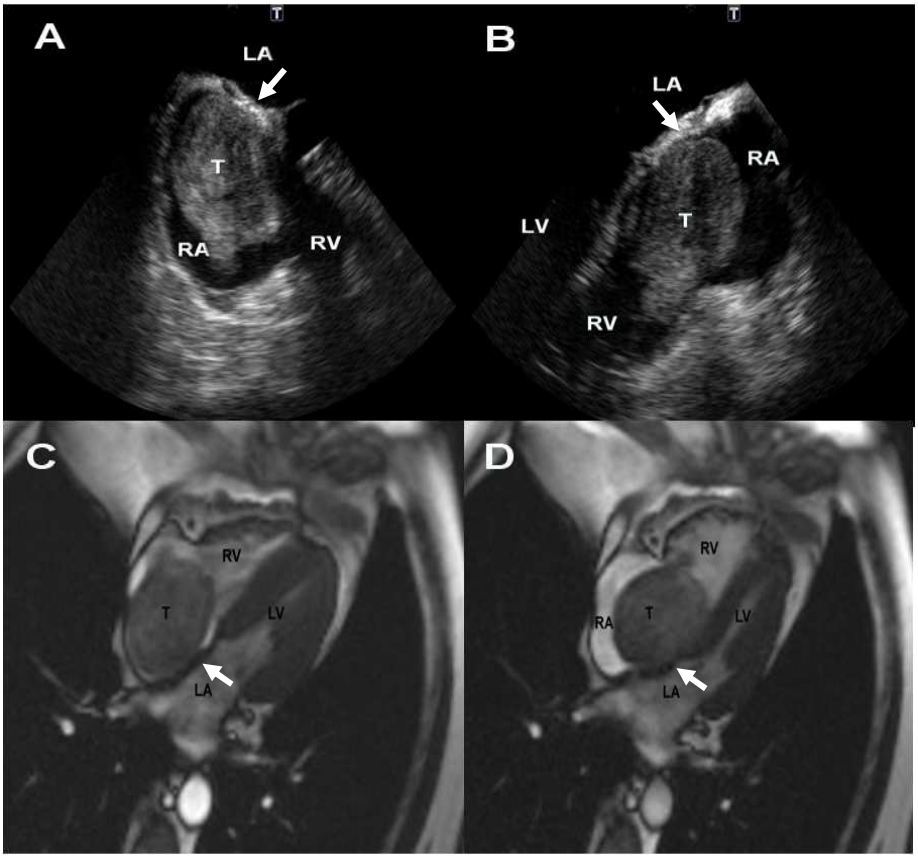
A 63-year-old man without history of cardiovascular diseases and without cardiovascular risk factors was referred to the Department of Cardiology because of extreme weight loss (about 20 kg within 6 months), weakness, fatigue, exertional dyspnea, and periods of hacking cough with no proper etiology. He was previously extensively examined (esophagogastroduodenoscopy, colonoscopy, bronchoscopy, and bone marrow biopsy) due to the commonest reasons for the mentioned symptoms – occult malignancy, hematological, autoimmune, and infectious diseases. Performed procedures didn't disclose any pathological substrate.

On physical examination, the patient was afebrile, eupneic at rest, and seriously emaciated (BMI 18.2 kg/m^2 compared to 12 months earlier BMI 24.4 kg/m^2). Chest auscultation was unremarkable. Cardiac auscultation revealed a regular rhythm of approximately 100 beats per minute, quiet heart sounds, and no murmurs or extra heart sounds. His arterial blood pressure was 130/90 mm Hg. There were no lower extremity edemas.

Electrocardiogram documented sinus tachycardia (100/min), right electrical axis, and left posterior hemiblock. Chest radiography showed no evidence of active lung lesions and was unremarkable in the context of cardiac pathology. Laboratory data revealed high erythrocyte sedimentation rate (110 mm/h) and C-reactive protein value (45.6 mg/L). All other hematological and biochemical parameters and tumor markers were within normal limits. Serum protein electrophoresis showed a slightly lower fraction of albumin (36.4 g/L), and insignificantly higher fraction of α-1 (5.3 g/L) and α-2 (13.2 g/L) proteins. There were no other deviations in the electrophoresis components.

Since extensive diagnostic evaluation performed during the first hospitalization showed no discrepancies, based on the above-mentioned laboratory (especially immunological) abnormalities, we suspected a heart tumor, primarily cardiac myxoma.

Transthoracic (TTE) and transesophageal echocardiography (TEE) revealed a large mobile tumor mass in the right atrium (approximately 57 x 41 mm), with the wide base attached to the interatrial septum, near the fossa ovalis (Picture 1 A and B).



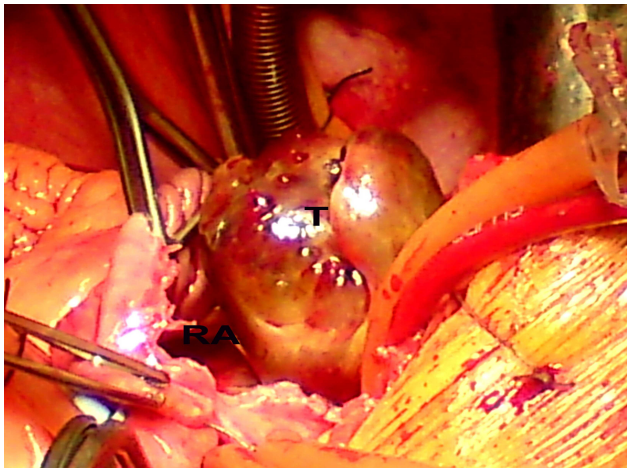
Picture 1 Transesophageal echocardiography (A and B) and cardiac magnetic resonance imaging (C and D) revealed a large solid, well-defined, expansive growth, measuring 57 x 41 mm in size, with a 21 mm base (arrow) attached to the atrial septum in the fossa ovalis segment. The tumor had almost completely filled the right atrium during systole (A and C), and prolapsed through the tricuspide valve during diastole causing relative tricuspide stenosis (B and D).

Slika 1. Ekokardiografija preko jednjaka (A i B) te kardijalna magnetska rezonanca (C i D) otkrivaju široku, dobro profiliranu i proširenu izraslinu veličine 57 x 41 mm s bazom od 21 mm (strelica) pričvršćena atrijskom septumu u segment fosse ovalis. Tumor je skoro u cijelosti popunio desnu preklijetku za vrijeme sistole (A i C) te spustio se u trolisni zalistak za vrijeme dijastole prouzrokujući relativnu trikuspidalnu stenozu (B i D).

RA – right atrium; RV – right ventricle; LA – left atrium; LV – left ventricle; T – tumor
RA – desna preklijetka; RV – desna komora; LA – lijeva preklijetka; LV – lijeva komora; T – tumor

The tumor extended during diastole through the tricuspid valve into the right ventricle causing relative tricuspid valve stenosis. Other echocardiographic parameters were unremarkable. Cardiac magnetic resonance imaging (CMRI) disclosed right atrial growth and confirmed the diagnosis of cardiac tumor (Picture 1 C and D). Multi-slice computed tomography (MSCT) angiography showed normal coronary arteries.

The tumor was completely surgically removed. It was a well-defined soft, encapsulated, brown-grey mass, with a smooth contour, sized 55 x 45 x 30 mm (Picture 2). The histology of the specimen revealed a cardiac myxoma (Picture 3).



Picture 2 Intraoperative view: the tumor (T) was a well-defined encapsulated mass with a smooth contour, brown colorized; almost completely filling the right atrium (RA).
Slika 2. Intraoperativni pregled: tumor (T) dobro profilirane enkapsulirane mase s glatkom konturom, smeđe boje, te gotovo u cijelosti popunjava desnu preklijetku (RA)

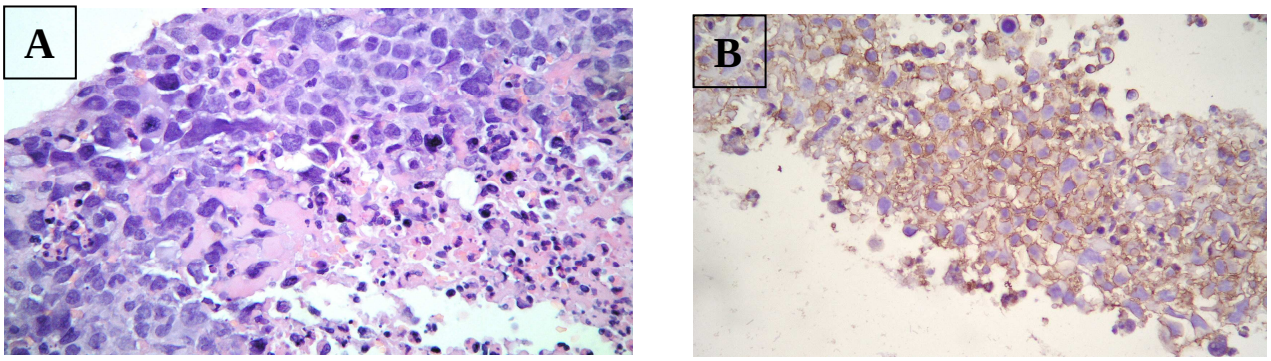
The patient was discharged uneventfully on 8th postoperative day. Five months later, the patient is well, with normal echocardiographic and laboratory findings, and increase in body weight by 15 kg.

Discussion

Myxoma is the most common cardiac tumor.[1,2] About 75% of cardiac myxomas arise in the left atrium, most frequently along the interatrial septum near the fossa ovalis.[1,2] Right atrial myxomas are estimated to occur in only 15-20% of the cases.[1,2] Sporadically, myxomas are located in the ventricules, valves, or in more than one heart chamber.[1,2] Although any age group can be affected, it predominates in the age group of 30-60 years with more than 75% of the affected being women.[1] Atrial myxomas mostly occur sporadically, but at least 7-10% of cases occur in a familial pattern, with an autosomal dominant transmission (e.g. Carney complex).[1,2]

Clinical presentations of cardiac myxomas mostly depend on tumor size, location, and mobility. Therefore, patients have various features of the classical Goodwin's myxoma triad consisting of cardiac (60%), embolic (30-40%), and constitutional symptoms (10-45%).[1,2]

Mechanical outflow tract obstruction may occur at the opening of any valve imitating the clinical presentation of valve stenosis.[3] Within that context, this can result in syncope or lethal valve obstruction.[1,4] However, the commonest obstructive symptom is exertional dyspnea that occurs in about 80% of patients with atrial myxoma.[1-3]



Picture 3 Histological findings supported the diagnosis of myxoma. The tumor consisted of a hypocellular mass of a myxoid matrix, rich in acid mucolike, elongated or stellate cells scattered in an abundant stroma (A, HE x 40; B, LCA).
Slika 3. *Histološki nalazi koji potvrđuju dijagnozu miksoma. Tumor je bio sastavljen od hipostanične mase miksoidne matice, bogate mukolitičnim produženim ili zvjezdastim ćelijama, razbacanima u obilnoj stromi (A, HE x 40; B, LCA).*

Cardiac signs and symptoms also include orthostatic hypotension, orthopnea, acute pulmonary edema, conduction alterations and disturbances, congestive heart failure, and sudden cardiac death.[1-4] Symptoms may manifest suddenly or intermittently, and can depend on body posture.[1,2]

Embolic phenomenon in cardiac myxoma is frequent with an estimated incidence between 30-40%.[1,2] Left-sided cardiac myxomas may be presented with embolization to the systemic circulation and cause specific clinical presentations depending on the affected organ.[1,2] Right-sided cardiac myxomas may remain asymptomatic or eventually cause pulmonary embolism due to tumor fragments or thrombus from the tumor surface, resulting in dyspnea, chest pain, hemoptysis, pulmonary hypertension, right-sided congestive heart failure, or sudden cardiac death.[1,2,4]

General or constitutional signs and symptoms (fatigue, weakness, weight loss, fever, arthralgia, myalgia, Raynaud's phenomenon, digital clubbing, erythematosus rash), as well as laboratory abnormalities (anemia, elevated erythrocyte sedimentation rate, serum C reactive protein and globulin levels, elevated leukocyte count, decreased platelet count, positive seroreactive protein) resembling connective tissue or malignant diseases were reported in 10-45% of patients.[1,2,5] It has been known that constitutional symptoms and laboratory findings are most often in patients with left atrial myxoma.[1] However, our case serves to remind that these symptoms can also be found in patients with a right-sided myxoma. The mentioned features are resolved immediately after surgery and are believed to be due to the release of inflammatory mediators from tumor cells, especially cytokine interleukine-6.[1,2,5]

Cardiac myxomas vary widely in size, and very little is known about their growth rate. It has been estimated that recurrent atrial myxomas grow an average of 0.24-1.6 cm per year.[1] Therefore, although symptoms occurred within 6 months before admission, the tumor's size in our patient assumed that intracardiac mass growth in a minimum of two to three years.

Findings on chest radiography of myxoma may be absent in one third of patients.[1,2] Calcification on chest radiography is more diagnostic of the right atrial myxoma, but is rarely present in the left atrial myxoma.[1,2] Transthoracic echocardiography (TTE) has approximately 90% sensitivity in the detection of cardiac myxoma, while the sensitivity of transesophageal echocardiography (TEE) is about 95%.[6,7] Specifically, it yields morphologic detail in the evaluation of cardiac tumors, including points of tumor attachment and degree of mobility.[6] TEE is also preferred because of its ability to detect other cardioembolic sources, such as intracardiac thrombus, vegetations, or aortic arch plaque.[7,8] MSCT and CMRI can help delineate the extent of the tumor and its relationships to the surrounding cardiac and thoracic structures.[9] Recently, 3-D TTE and TEE have emerged as a novel and useful tool for the detection and differential diagnosis of intracardiac growths.[4,7,8,10] Nevertheless, differential diagnosis may be difficult until surgical excision and histological examination are performed.[8]

To summarize, although clinical presentation of cardiac myxoma is well known, our experience confirms that the diagnosis of this tumor usually begins after the exclusion of other pathology or after the occurrence of cardiac symptoms.[3,4] This case reminds us that cardiac myxoma should be considered as a possible etiology of extensive weight loss, weakness and laboratory abnormalities. Therefore, we suggest that echocardiography, as a widely available, noninvasive method, should be performed early in the diagnostic evaluation of these patients in order to detect rare but treatable heart tumors.

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